Lingual thyroid & its manage-ment

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Lingual thyroid is a rare condition. It is seen roughly 1 in 100,000 populations. Managing this condition is filled with historical controversies ranging from leaving it alone to surgical removal of the lesion. Attempt has been made to present in a precise way the management modalities available. All the surgical modalities along with their pluses and minuses are discussed here.

11/7/2010

Introduction:

Lingual thyroid is caused by a rare developmental disorder caused due to aberrant embryogenesis during the descent of thyroid gland to the neck. Lingual thyroid is the most frequent ectopic location of thyroid gland. Prevalence rates of lingual thyroid vary from 1 in 100,000 to 1 in 300,000. Review of literature reveals that only about 400 symptomatic cases have been reported so far. This could well be an understatement and statistical anomaly.

History:

Hickmann recorded the first case of lingual thyroid in 1869. Montgomery stressed that for a condition to be branded as lingual thyroid, thyroid follicles should be demonstrated histopathologically in tissues sampled from the lesion.

Common locations of ectopic thyroid gland include:

1. Between geniohyoid and mylohyoid muscles (sublingual thyroid)
2. Above the hyoid bone (suprahyoid prelaryngeal)
3. Mediastimum
4. Pericardial sac
5. Heart
6. Breast
7. Pharynx
8. Oesophagus
9. Trachea
10. Lung
11. Duodenum
12. Mesentery of small intestine
13. Adrenal gland

Embryology:
A brief discussion of embryology of thyroid gland will not be out of place as this would ensure better understanding of the pathophysiology involved in the formation of ectopic thyoid gland.
Initially thyroid gland appears as proliferation of endodermal tissue in the floor of the pharynx between tuberculum impar and hypobranchial eminence (this area is the later foramen caecum). Cells of thyroid gland descend into the mesoderm above aortic sac into the hypopharyngeal eminence (later pharynx) as cords of cells. During this descent thyroid tissue retains its communication with foramen cecum. This communication is known as thyroglossal duct. This duct disappears as soon as the

descent is complete.
Thyroid gland descends in front of the hyoid bone and laryngeal cartilages. By 7th week it reaches its final destination in front of trachea. At this time a small median isthmus develops connecting the lobes of thyroid gland. The gland begins to function by the 3rd month when thyroid follicles start to develop. Parafollicular or c cells that secrete calcitonin are developed from ultimobranchial bodies.

Persistence of thyroglossal duct even after birth leads to the formation of thyroglossal cyst. These cysts usually arise from the remnants of thyroglossal duct and can be found anywhere along the migration site of thyroid gland. They are commonly found behind the arch of hyoid bone. Important diagnostic feature is their midline location.

Normal development and migration of thyroid gland needs an intact Tbx1-Fgf8 pathway. This pathway has been identified as the key regulator of development of human thyroid gland. Tbx1 regulates the expression of Fgf8 in the mesoderm, it is postulated that Fgf8 mediates critical Tbx1-dependent interactions between mesodermal cells and endodermal thyrocyte progenitors.
Tbx1 is not expressed by thyroid primordium, but is strongly expressed by the surrounding mesoderm. It is also expressed by pharyngeal endoderm lateral to thyroid primordium.
Thyroid organogenesis associated with the expression of a set of transcription factor encoding genes. They include Nkx2-1, Foxe1, Pax8 and Hhex1 genes. Expression of these genes in thyroid primordium is also dependent on Tbx1 gene expression.

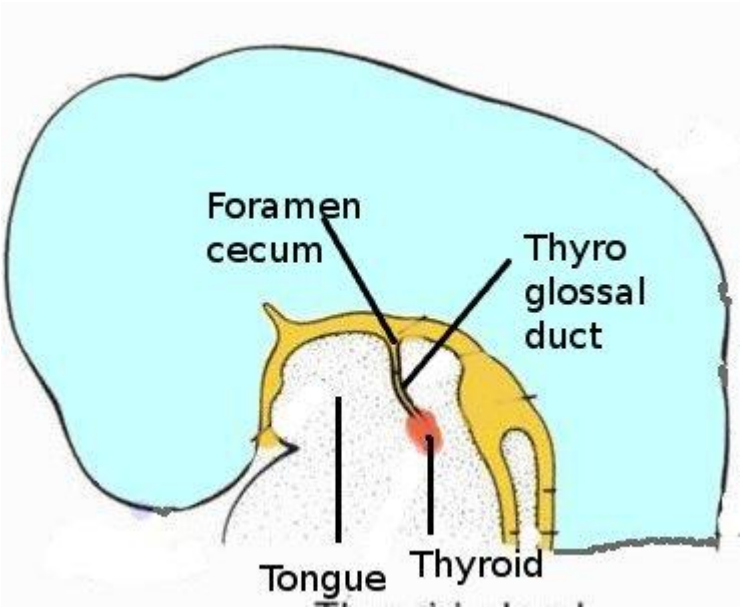


Figure showing development of thyriod ventral to foramen cecum

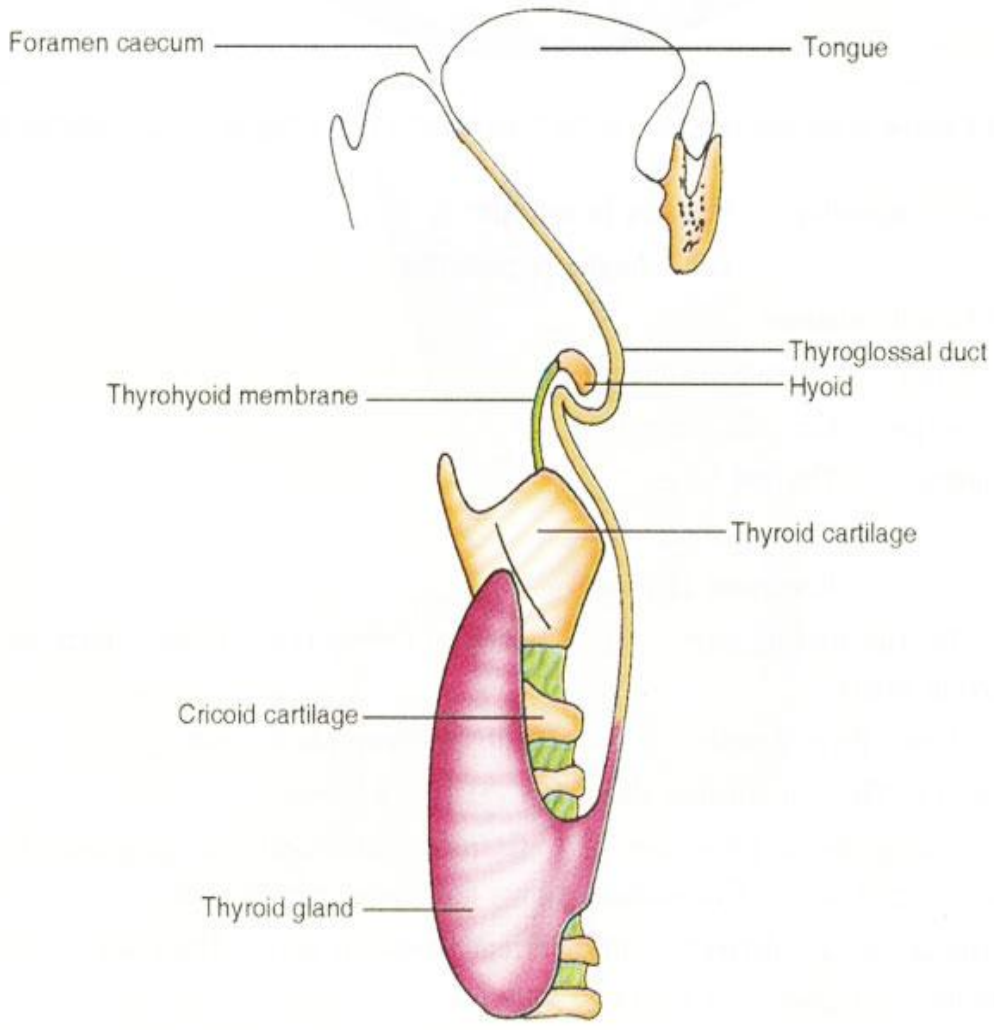


Figure showing migration of thyroid gland

It commonly occurs in females. Female:Male ratio is 4:1. Even though lingual thyroid may manifest at any age it is commonly seen in patients in whom there is extra demand of thyroxin by the body which causes it to undergo physiological enlargement. It is commonly seen during early childhood and teens.

Symptoms:

Majority of these patients are asymptomatic. They will have no problems other than swelling in the posterior portion of their tongue.
Symptoms caused by lingual thyroid include:

1. Dysphagia
2. Dysphonia
3. Bleeding from the mass
4. Sleep apnoea
5. Hypothyroidism
6. Dyspnoea (rarely)

In rare cases lingual thyroid could undergo malignant transformation.

Features seen on examination:

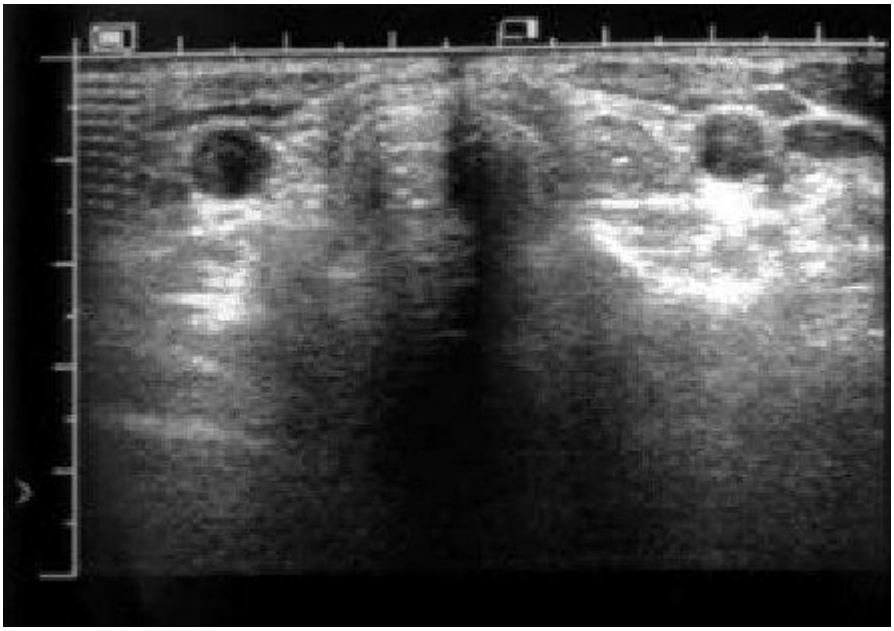
Clinical photograph showing lingual thyroid mass

Lingual thyroid could be seen as pinkish mucosa covered mass over the posterior third of tongue. On palpation this mass could be felt as solid firm and fixed mass. It would be seen attached to the tongue at the junction of anterior 2/3 and posterior 1/3. This is where approximately foramen cecum is supposed to be present. Attempt should be made to palpate the neck in the region of thyroid to ascertain whether normal thyroid tissue is present in the neck.

Investigation:

Ultrasound neck:

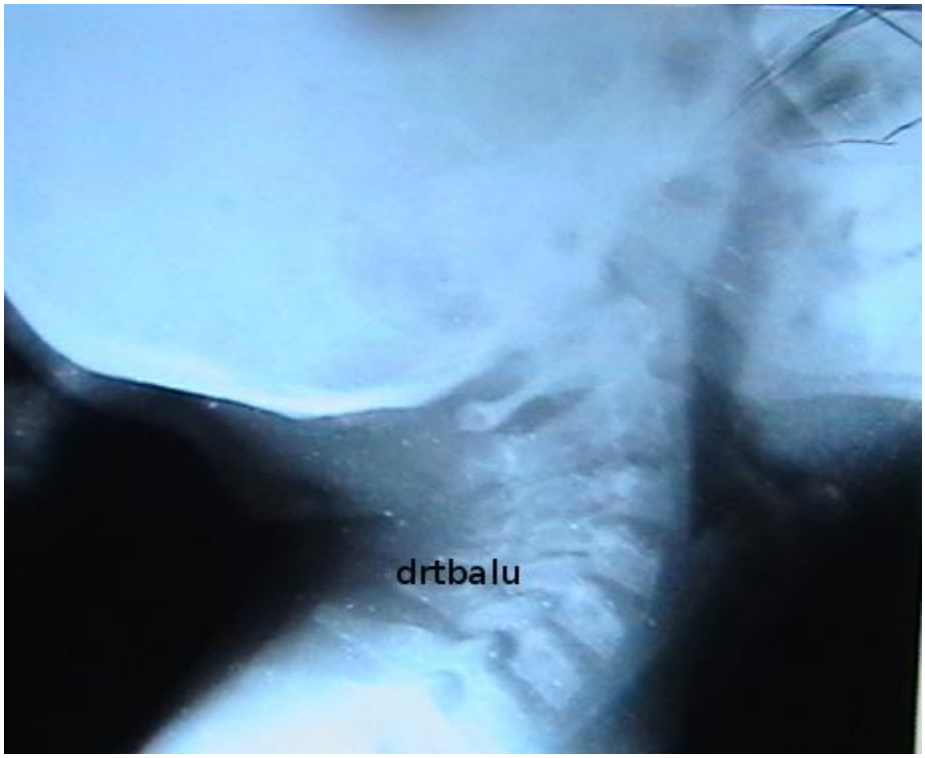
In all patients with lingual thyroid the presence of normal thyroid in the neck should be ascertained. This can easily be done by performing ultrasound examination of neck. It will reveal the presence or absence of normal thyroid gland in the neck.



Picture showing ultrasound neck with absence of thyroid gland in the neck

X-ray soft tissue neck lateral view:

This will just reveal the presence of soft tissue shadow in the region of the tongue. It will also demonstrate the lower extent of the mass.

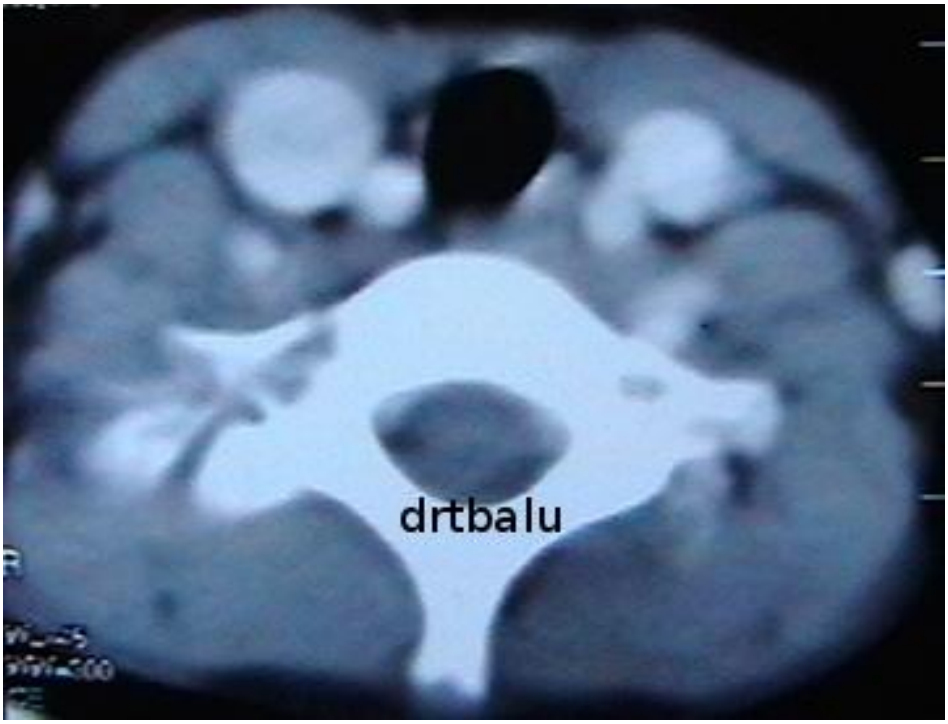


X-ray soft tissue neck lateral view showing a globular soft tissue mass in the region of tongue above the level of hyoid bone

CT scan:

This will help in accurately assessing the extent of lesion. If contrast is used it would give valuable input regarding its vascularity. CT scan of neck will also categorically reveal the presence or absence of normal thyroid tissue in the neck.

CT scan axial cut taken at the level of lower border of mandible clearly shows soft tissue mass occupying the posterior portion of tongue.



CT scan neck axial view with contrast shows absence of thyroid gland in the neck. The internal jugular vein and carotid artery could be seen as enhancing masses. Jugular vein of one side appears to be predominantly enlarged.

Technitium 99 scan is virtually diagnostic. It will clearly reveal the radioactive isotope uptake by the thyroid tissue present on the tongue. It will also clearly demonstrate the presence or absence of thyroid tissue in the neck region. These images are obtained in either dynamic or static mode 20 minutes after intravenous injection of 74-111MBq of Technitium 99 pertechnetate. Its molecular weight is comparable to that of iodine and is transported actively into the thyroid tissue via the sodium iodide symporter system.

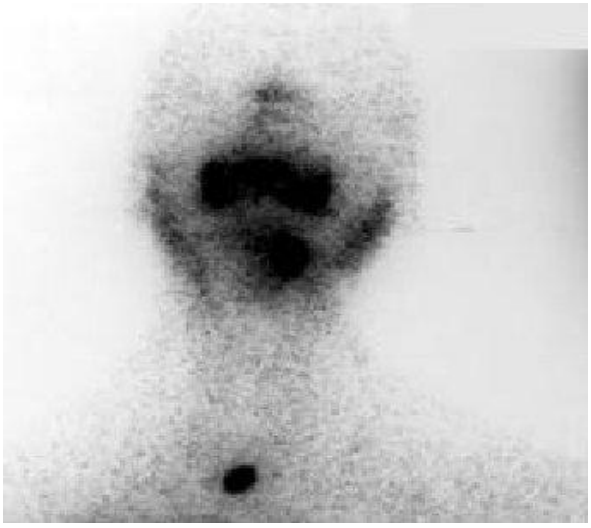


Figure showing Technitium 99 scan. It clearly shows increased uptake in the region of the tongue (due to lingual thyroid tissue) and absence of uptake in the neck region due to absence of normal thyroid tissue in this area.

Role of radio active iodine uptake studies:

This helps in ascertaining the functional status of the thyroid gland. It also helps in ascertaining the viability of the transplanted extopic thyoid gland 100 days after the surgical procedure.
Both I 131 and I 123 can be used for this purpose. I 123 has a favourable dosimetry for imaging. Since it is produced in a cyclotron it is rather expensive. Whereas I 131 is reactor produced and is reasonable cheap. It is also freely available. It has poor imaging characteristics and emits beta radiation. Its half life is about 8 – 10 days as compared to 12 hours of I 123. Hence I 123 is preferred for functioning radioactive imaging purposes.
Radioactive iodine is usually administered in small doses orally and uptake is measured at different intervals i.e. 2 hrs, 4 hrs, 24hrs and 48 hrs.

Estimation of serum T3 T4 and TSH levels:

This will help in assessing the functional status of the ectopic gland. Invariably majority of these patients are euthyroid. If TSH levels are raised then suppression

can be attempted using regular doses of oral thyroxine.

Management:

Conservative: If the lingual thyroid is the only functioning thyroid suppression therapy using regular oral doses of thyroxine can be attempted. This is more so in patients whose normal physiological requirement of thyroxine is raised as during periods of active growth, menarche, pregnancy etc. This suppression therapy will help in preventing abnormal physiological enlargement of the ectopic thyroid tissue.

Surgical management:

Indications for surgery:

1. If the mass produces obstructive symptoms
2. If the mass produces bleeding
3. If the mass demonstrates sudden increase in size
4. If malignancy is suspected

FNAC is not advised as it would cause unnecessary bleeding. Similarly instead of biopsying the lesion total excision is preferred.

Methods of excision:

Transoral method of excision:

This method of excision is preferred for small lingual thyroid masses. It is ideally suited for lesions which are above the level of hyoid bone. Clinically if the posterior border of the swelling is seen on depressing the tongue with a tongue depressor then one can safely go ahead and remove the mass transorally.
Transoral removal is assisted by:

1. Cautery
2. Coablation
3. Debrider
4. Laser

Surgery is usually performed under general anesthesia induced via nasotracheal intubation. This is the preferred intubation modality in these patients as it would avoid troublesome bleeding following intubation trauma.
Patient is placed in Rose position. Boyles Davis mouth gag is used to hold the mouth open. Throat is packed tightly using ribbon gauze to avoid spillage into larynx. The

mass is held with a tenaculum forceps and is pulled anteriorly. The anterior border is incised using diathermy cautery / coblator /laser. The tumor is gently dissected and stripped away from the lingual tissue. Perfect hemostasis is secured by coagulating the bleeding points seen in the base of the tumor.

Debrider blade can be used to shave off the tumor from the tongue base. Bleeding points seen in the base can be cauterized using bipolar cautery.

Advantages of transoral approach:

1. Easy to perform
2. Neck incision is avoided
3. Patient's recovery is rapid
4. Complications are minimal

Transmandibular translingual approach:

This approach is very useful in removing very large lingual thyroid masses.

Procedure:

Preliminary tracheostomy is performed under local anesthesia. General anesthesia is introduced via tracheostome. This protects and takes control of the airway in an efficient manner.
An incision over the mucoperiosteum of the buccogingival sulcus is performed over the interior region of mandible and the bone over the mental area is exposed. A midline vertical osteotomy of the mandible is performed. The tongue is sectioned sagittally in the midline up to the floor of the mouth till the tongue base is reached. The lingual thyroid mass is excised in toto. The wound is closed in layers. The mandible is immobilized by wiring and dental arch bar.

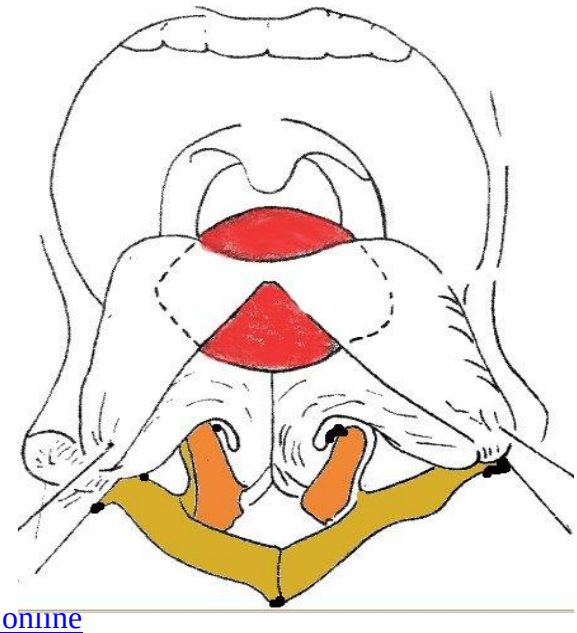


Figure showing the transmandibular approach

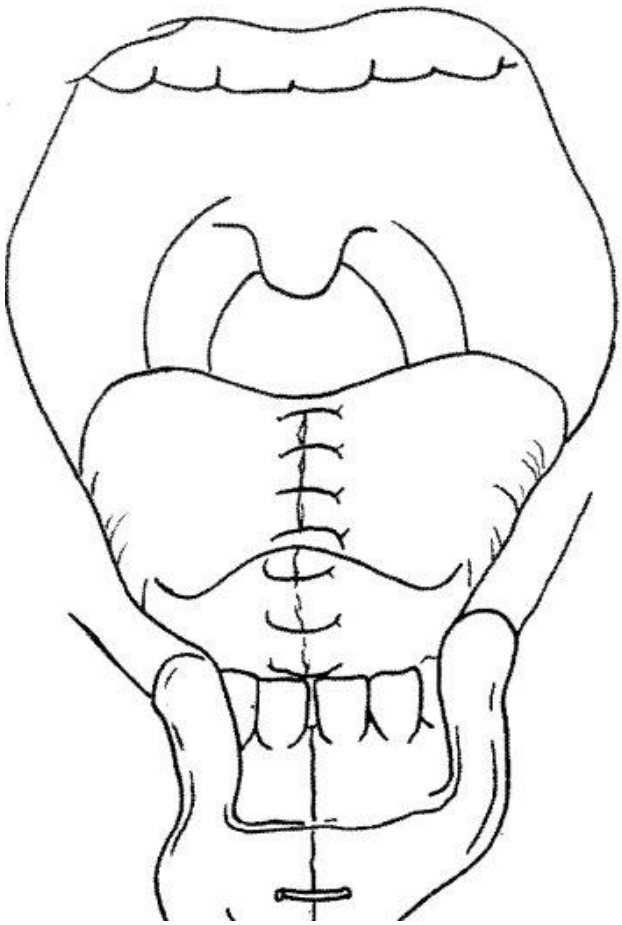


Figure showing wound closure

Advantages:

1. Excellent visualization
2. No need for ligating lingual vessels
3. Important structures are spared i.e lingual nerve, hypoglossal nerve, and submandibular salivary gland

Lateral pharyngotomy approach:

This approach is preferred if transpositioning of lingual thyroid is planned. Anaesthesia is induced via nasotracheal intubation. Patient is positioned in such a way that the neck is slightly extended. An oblique curved incision is made about 8

cms long in the left lateral portion of upper neck just anterior to sternomastoid muscle. The dissection is proceeded in the subplatysmal plane. The following structures are identified:

1. Carotid bifurcation
2. Lingual artery
3. Superior thyroid artery
4. Hypoglossal nerve

Using the finger guide passing through the oral cavity to the left lateral pharynx at the level of base of tongue a lateral transverse pharyngotomy of 3-4 cms is made inferior to the hypoglossal nerve and above the hyoid bone.
Through this pharyngotomy opening the posterior 1/3 of tongue, epiglottis and lingual thyroid mass could be identified. The gland is dissected out of the tongue. The right side of the mass is totally freed of the tongue. The mass is mobilised by an encircling incision over the tongue. A small attachment to the left side of tongue base is retained. This will ensure adequate vascularity to the mass after transposition. The mass is delivered via the pharyngotomy opening and is implanted in the left side of the neck with its attachment to the left tongue base remaining intact. The wound is closed in layers.

Advantage:

The most important advantage of this approach is that it ensures tension free transposition of lingual thyroid to the left side of neck. After transposition the gland can easily be examined on the left lateral neck of the patient.

Suprahyoid midline approach:

This approach is preferred in removing large lingual thyroid mass even if it extends to a level below that of hyoid bone.

Procedure:

This surgery is performed under general anesthesia administered via nasotracheal intubation. This intubation modality prevents intubation injury to lingual thyroid mass.

Infiltration:

The surgical area in the neck is liberally infiltrated using Tumescent fluid.

Tumescent fluid is prepared using:

1. one litre of ringer lactate solution
2. 40 ml of 2% xylocaine
3. 1ml of 1 in 1000 adrenaline
4. 20 ml of 8.4% soda bicarb

Advantages of using Tumescent fluid infiltration:

1. Breaks open tissue planes facilitating easy dissection i.e Hydrodissection
2. Reduces bleeding due to vasoconstrictive effect of adrenaline
3. Facilitates uniform heat dissipation when diathermy is used during surgical procedure
4. Prevents development of local tissue level acidosis

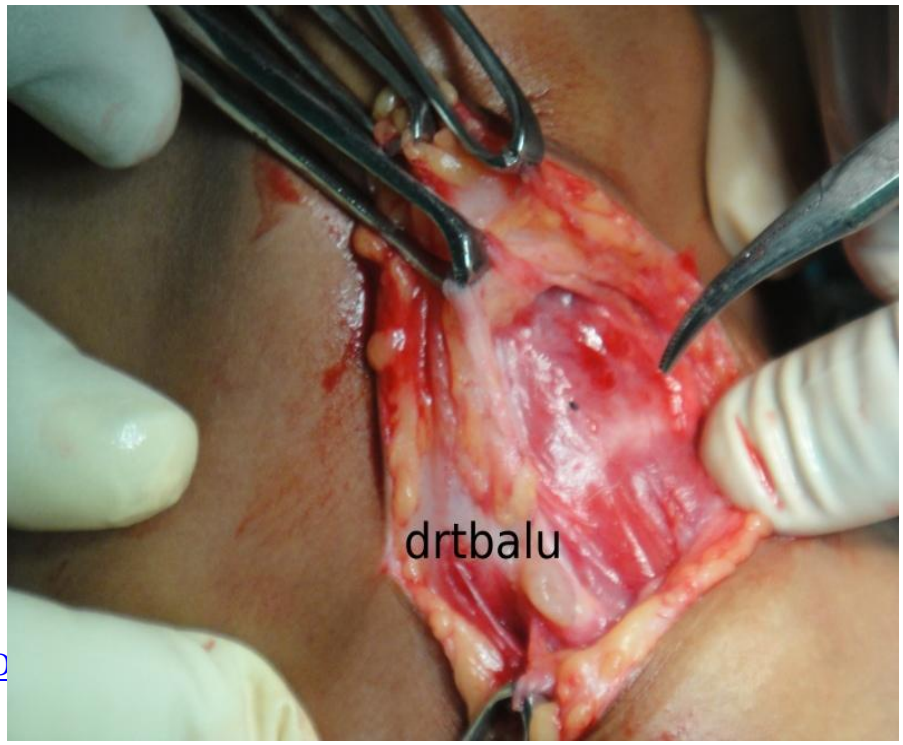
Figure showing infiltration being given

Incision:

Transverse skin crease incision is made at the level of hyoid bone. Skin, subcutaneous tissue and cervical fascia are elevated in the subplatysmal plane. Sticking on to the subplatysmal plane helps in preserving the cervical branches of

facial nerve. Dissection in this plane is continued and the flap is raised above the level of hyoid bone.

Incision being widened using cutting diathermy



Hyoid bone visualized

Supra hyoid dissection:

In this stage the muscles attached to the hyiod bone are cut and dissected subperiosteally.

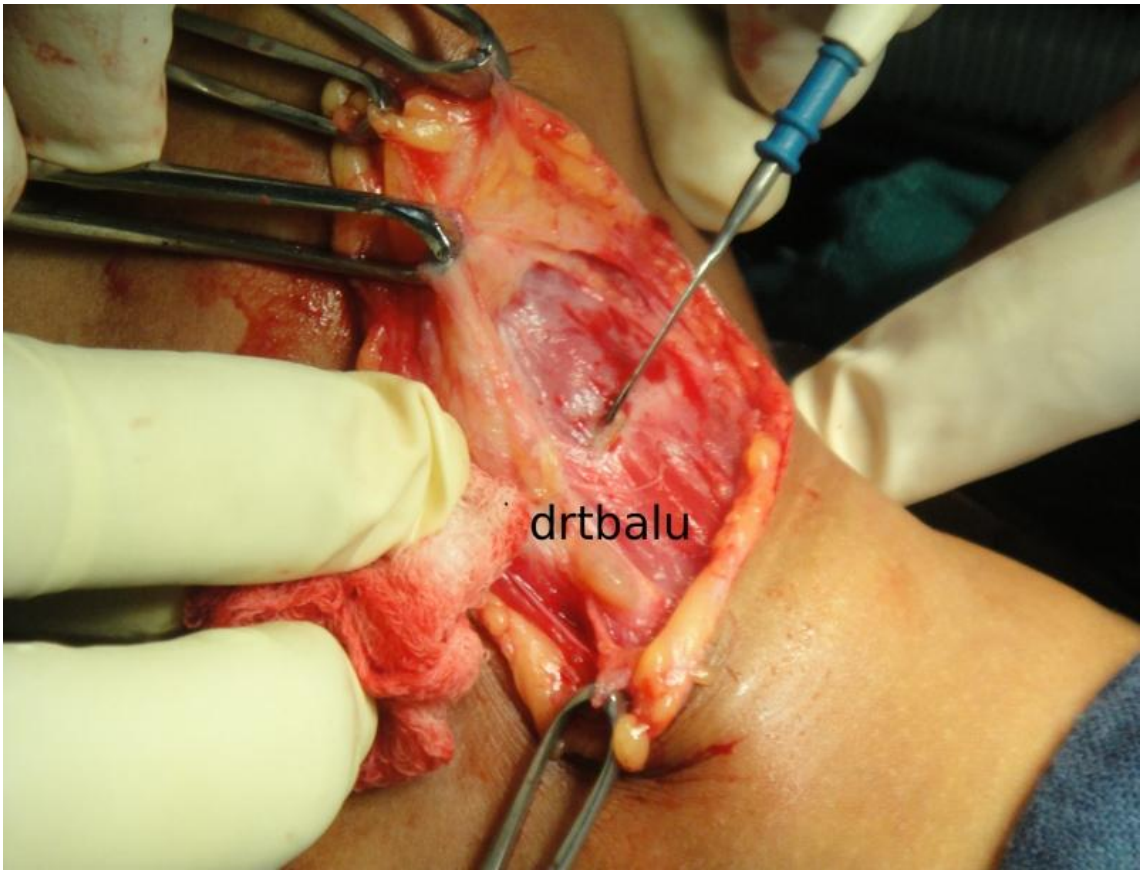


Figure showing hyoid bone being skeletonized using a cutting diathermy

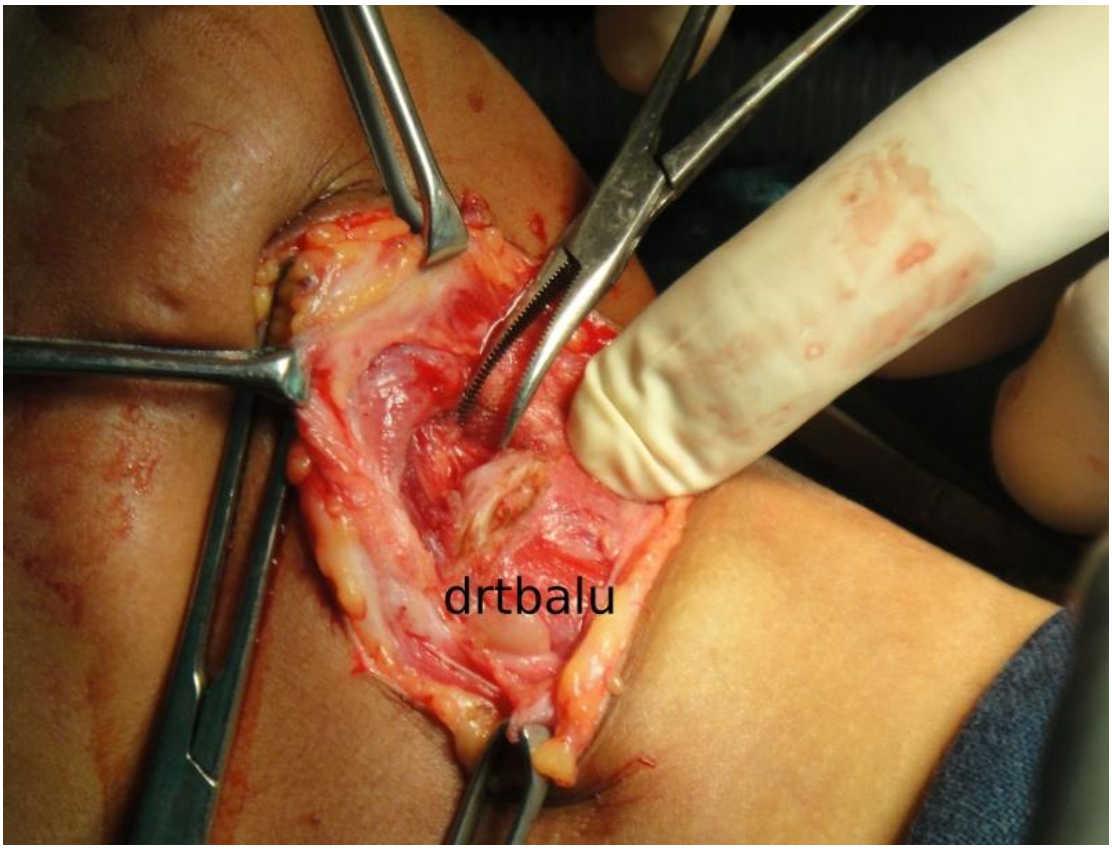


Figure showing suprahyoid subperichondrial dissection being performed

The supra hyoid muscles are split and the oral cavity is entered. Using a finger guide inside the oral cavity the mass is pushed downwards and delivered via the suprahyoid neck incision. The mass is removed in full. The wound should be meticulously closed in layers. Ryles tube should be inserted to facilitate early feeding. Ideally the Ryles tube should be left in place at least for 3 days.

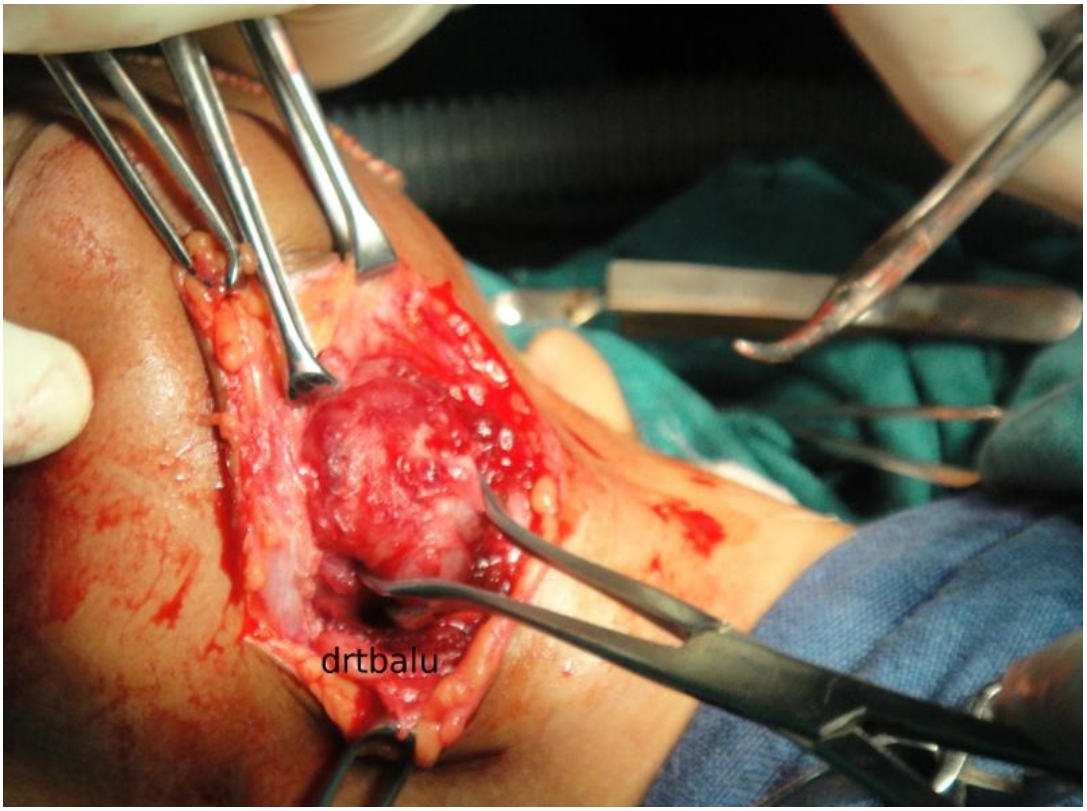
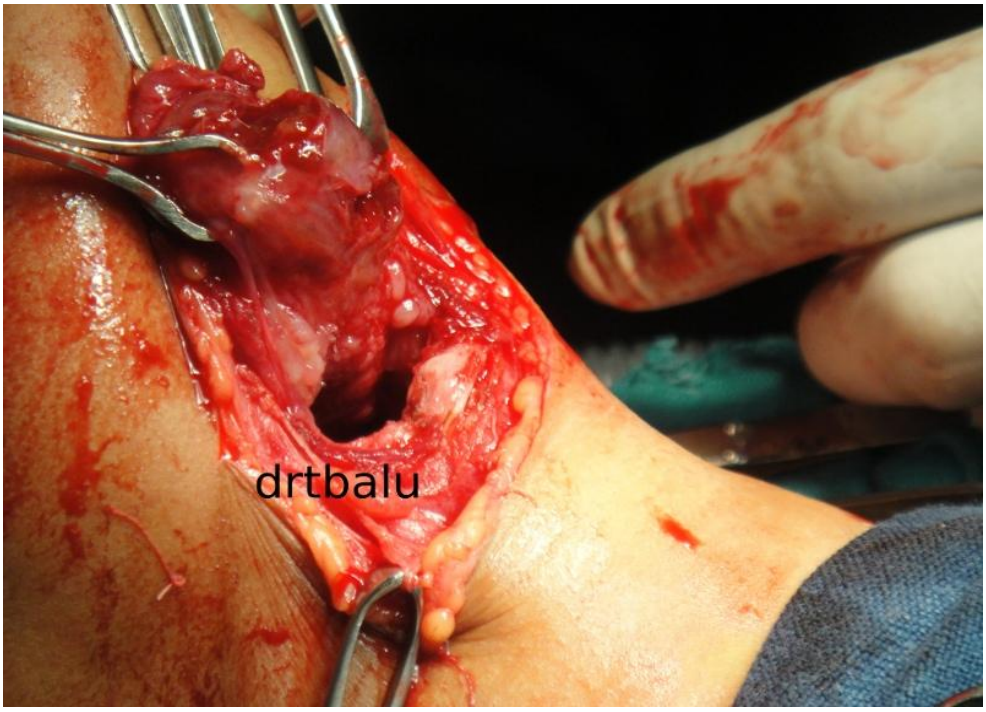


Figure showing lingual thyroid being delivered in to the neck



Lingual thyroid attached to the base of tongue

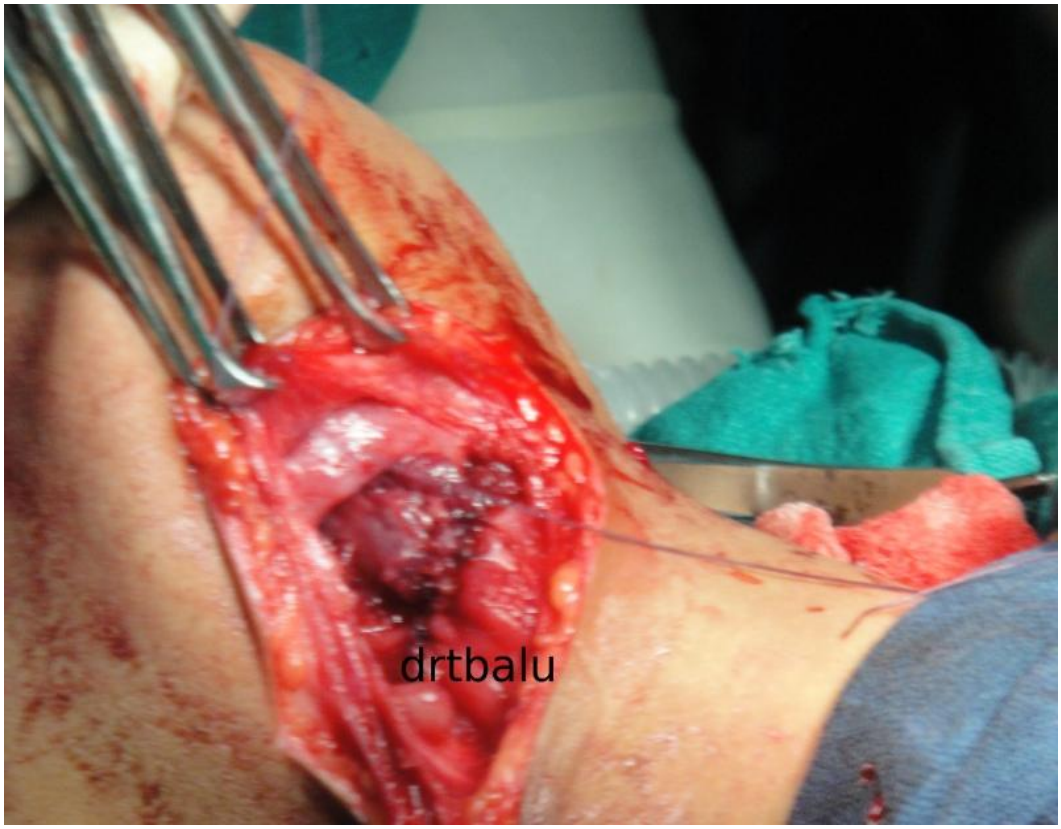


Figure showing wound closure

After surgery all these patients should be started on oral supplemental doses of thyroxine.

If you are wondering about the status of parathyroids, you need not worry as they will be in their normal position i.e. neck because embryologically their developmental process is different.